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PART XXVI The Endocrine System

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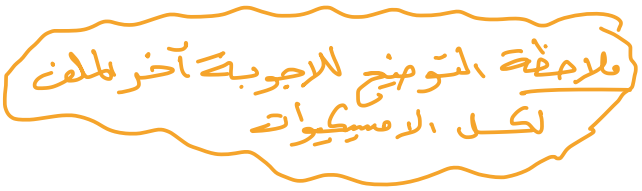
1. Main causes of hypocalcemia in nephrotic syndrome in children are the following **EXCEPT**
   A. decreased activity of renal α-hydroxylase
   B. lowered serum albumin
   C. decreased intestinal absorption of calcium
   D. increased urinary losses of cholecalciferol-binding globulin
   E. prednisone therapy

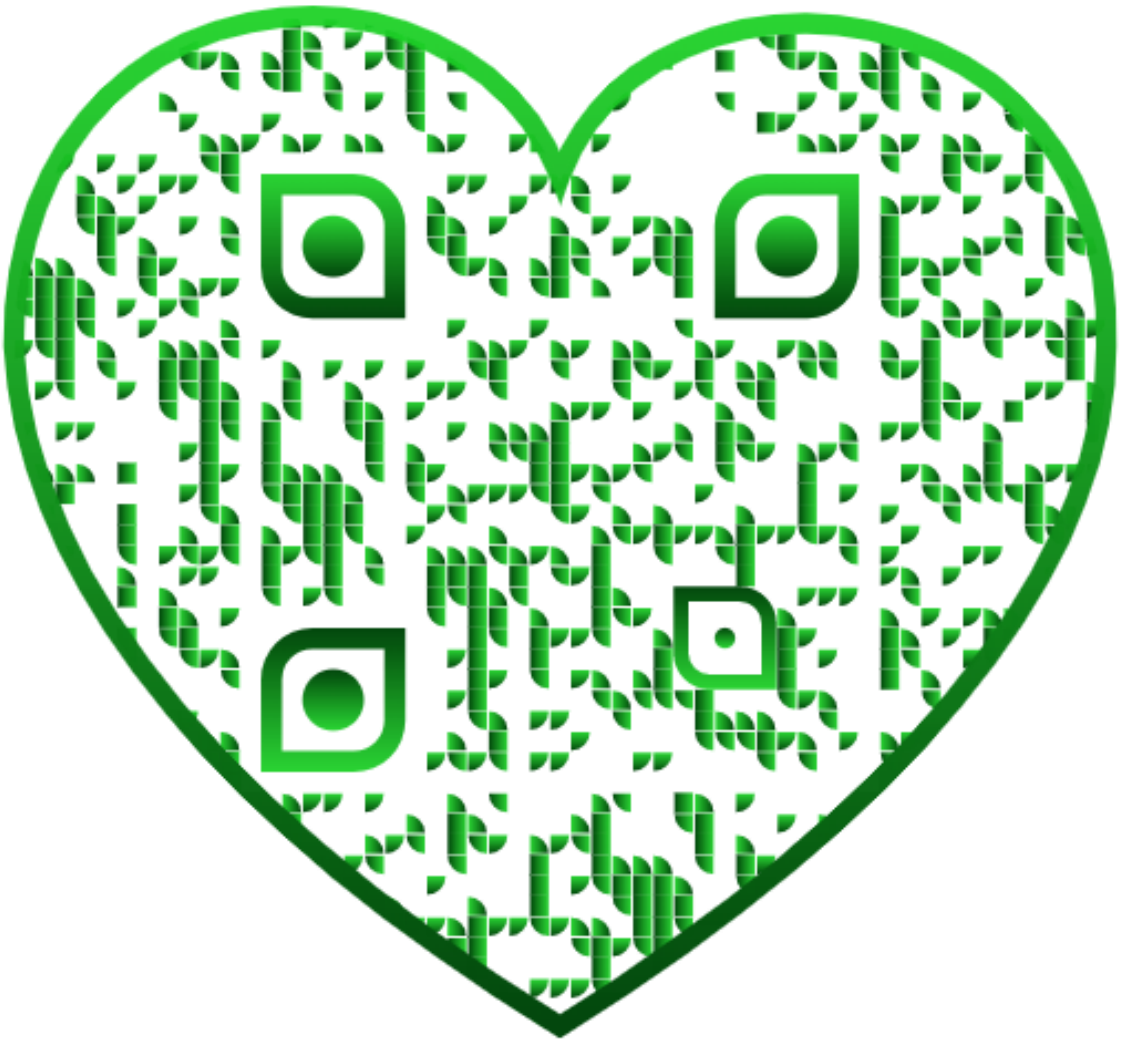
2. The following criteria are needed for the diagnosis of syndrome of inappropriate secretion of antidiuretic hormone (SIADH) **EXCEPT**
   A. excessive urinary sodium concentration
   B. hyponatremia with normal serum osmolality
   C. elevated urine osmolality
   D. normal renal, adrenal, and thyroid function
   E. absence of volume depletion

3. The following are main features of diabetic ketoacidosis (DKA) **EXCEPT**
   A. hyperglycemia (glucose usually >300 mg/dL)
   B. ketonemia (serum ketones >3 mmol/L)
   C. ketonuria
   D. venous pH <7.20
   E. serum HCO_3<15 mEq/L

4. What percent of newly diagnosed diabetics present with symptoms of diabetic ketoacidosis (DKA)?
   A. 10%
   B. 20%
   C. 30%
   D. 40%
   E. 50%

5. Typical insulin dosage requirement in prepubertal children after the "honeymoon" period is
   A. 0.4 U/kg/day
   B. 0.5 U/kg/day
   C. 0.6 U/kg/day





D. 0.7 U/kg/day
E. 1 U/kg/day

6. The following features suggest constitutional delay as a cause of short stature **EXCEPT**

A. no signs or symptoms of systemic disease
B. bone age delayed beyond the height age
C. period of poorest growth often occurring between the ages of 18 and 30 months
D. parental or sibling history of delayed development
E. height predictions consistent with family characteristics

7. Congenital goiter is seen in which percent of newborns with congenital hypothyroidism?

A. 10%
B. 20%
C. 30%
D. 40%
E. 50%

8. During a routine physical examination, a solitary thyroid nodule is palpated in asymptomatic 10-year-old child.
Of the following, the **MOST** likely cause is

A. thyroid carcinoma
B. thyroid adenoma
C. thyroid abscess
D. thyroid cyst
E. subacute thyroiditis

9. The pituitary gland is located in a saddle-shaped cavity of the

A. frontal bone
B. maxillary bone
C. sphenoid bone
D. ethemoid bone
E. temporal bone

10. Physiologic factors play a role in stimulating and inhibiting growth hormone (GH). One of the following inhibit GH release

A. sleep
B. exercise
C. hyperglycemia
D. acute illness

E. fasting

11. Corticotropin-releasing hormone (CRH) and ACTH release are inhibited by
A. arginine vasopressin
B. oxytocin
C. angiotensin II
D. atrial natriuretic peptide
E. cholecystokinin

12. The **MOST** common cause of acquired hypopituitarism is
A. craniopharyngioma
B. eosinophilic granuloma (histiocytosis)
C. tuberculosis
D. toxoplasmosis
E. meningitis

13. Indications for GH treatment to promote growth include the following **EXCEPT**
A. GH deficiency
B. chronic renal failure before transplantation
C. celiac disease
D. Prader-Willi syndrome
E. Noonan syndrome

14. Reported side effects of GH treatment include the following **EXCEPT**
A. pseudotumor cerebri
B. slipped capital femoral epiphysis
C. gynecomastia
D. worsening of scoliosis
E. 6-fold increase in the risk for type 1 diabetes

15. The following stimulate vasopressin (VP) secretion **EXCEPT**
A. hyperosmolality
B. hypovolemia
C. hypotension
D. hyperglycemia
E. nausea

16. Acquired nephrogenic diabetes insipidus (NDI) is associated with the following **EXCEPT**
A. lithium
B. vancomycin
C. amphotericin

D. methicillin
E. rifampin

17. Neonates and young infants with central diabetes insipidus are often best treated with
A. fluid therapy
B. vasopressin analogs
C. thiazide diuretics
D. indomethacin
E. amiloride

18. Hyponatremia in cerebral salt wasting is accompanied by the following **EXCEPT**
A. elevated urinary sodium excretion
B. low urine output
C. hypovolemia
D. normal or high uric acid
E. suppressed vasopressin

19. Broad forehead, hypertelorism, small chin, long philtrum, camptodactyly, and fetal finger pads are features of the following genetic overgrowth syndrome
A. Perlman syndrome
B. Sotos syndrome
C. Weaver syndrome
D. Beals syndrome
E. Beckwith-Wiedemann syndrome (BWS)

20. Precocious puberty in girls is defined by the onset of secondary sexual characteristics before the age of
A. 7 yr
B. 8 yr
C. 9 yr
D. 10 yr
E. 11 yr

21. The **MOST** common brain lesion causing central precocious puberty is
A. postencephalitic scar
B. tuberculous meningitis
C. hypothalamic hamartoma
D. tuberous sclerosis
E. hydrocephalus

22. All the following are true in premature thelarche **EXCEPT**

A. most often appears in the 1st 2 yr of life
B. growth and osseous maturation are usually normal
C. menarche occurs at the expected age
D. may be unilateral
E. often persists till puberty

23. Premature adrenarche has traditionally applied to the appearance of sexual hair before the age of 8 yr in girls or 9 yr in boys without other evidence of maturation. It is characterized by the following **EXCEPT**

A. more frequent in girls
B. higher prevalence in african-american
C. axillary hair generally appears earlier
D. adult-type axillary odor is common
E. affected children are often slightly advanced in height

24. The **MOST** accurate test of thyroid function is

A. T4
B. free T4
C. T3
D. thyroglobulin
E. TSH

25. Peak serum concentrations of TSH at birth in full-term infants reach

A. 20 mU/L
B. 40 mU/L
C. 60 mU/L
D. 80 mU/L
E. 100 mU/L

26. The thyroid hormones are transported in plasma bound to thyroxine-binding globulin (TBG), a glycoprotein synthesized in the liver. TBG binds approximately 70% of T4 and 50% of T3. TBG level increase with administration of

A. estrogens
B. androgens
C. glucocorticoids
D. nicotinic acid
E. l-asparaginase

27. The **MOST** common cause of permanent congenital hypothyroidism is

A. dyshormonogenesis
B. thyroid dysgenesis
C. Iodine deficiency

D. defect of iodide transport
E. Pendred syndrome

28. The cord serum T4 is decreased in proportion to gestational age and birthweight. The serum T4 gradually increases and enters the T4 range seen in term infants by the age of
A. 2 wk
B. 4 wk
C. 6 wk
D. 8 wk
E. 10 wk

29. Approximately 10% of infants with congenital hypothyroidism have associated congenital anomalies.
Of the following, the **MOST** common are
A. nervous system anomalies
B. lungs anomalies
C. eye anomalies
D. cardiac anomalies
E. genitourinary anomalies

30. The normal level of serum thyroid-stimulating hormone in first week of term Infants is up to
A. 4.4 mIU/L
B. 9.5 mIU/L
C. 13.6 mIU/L
D. 17.6 mIU/L
E. 27.0 mIU/L

31. Retardation of osseous development in congenitally hypothyroid infants can be shown radiographically at birth in approximately
A. 20%
B. 40%
C. 60%
D. 80%
E. 100%

32. Levothyroxine (l-T4) given orally is the treatment of choice in congenital hypothyroidism.
The recommended initial starting dose is
A. 2-5 µg/kg/day
B. 4-7 µg/kg/day

C. 5-9 µg/kg/day
D. 8-12 µg/kg/day
E. 10-15 µg/kg/day

33. The **MOST** common cause of acquired hypothyroidism is
A. craniopharyngioma
B. Hashimoto thyroiditis
C. meningoencephalitis
D. drug-induced
E. irradiation

34. Antithyroid antibodies develop and subclinical or overt hypothyroidism occurs in the following conditions **EXCEPT**
A. Down syndrome
B. Turner syndrome
C. type 1 diabetes mellitus
D. Sjögren syndrome
E. Williams syndrome

35. Risk for subclinical hypothyroidism is seen in children with
A. chronic hepatitis B infection
B. chronic hepatitis C infection
C. chronic hepatitis D infection
D. hepatitis A infection
E. hepatitis E infection

36. The first clinical manifestation of acquired hypothyroidism is
A. poor schoolwork
B. goiter
C. deceleration of growth
D. weight gain
E. constipation

37. One of the following is not routinely done in children with suspected hypothyroidism
A. sonography
B. serum free T4
C. TSH
D. antithyroglobulin
E. antiperoxidase antibodies

38. Chronic lymphocytic thyroiditis is the most common cause of acquired hypothyroidism, with or without goiter.
All the following are true **EXCEPT**

A. more common in girls
B. peak incidence during adolescence
C. most of the affected children are asymptomatic
D. clinical course is constant
E. familial clusters are common

39. Thyroid peroxidase antibodies are absent in the following thyroiditis syndrome

A. Hashimoto thyroiditis
B. painless sporadic thyroiditis
C. painful subacute thyroiditis
D. acute suppurative thyroiditis
E. Riedel thyroiditis

40. Endemic cretinism is the most serious consequence of iodine deficiency; it includes two different but overlapping syndromes: a neurologic type and a myxedematous type. The neurologic syndrome is characterized by the following **EXCEPT**

A. intellectual disability
B. deaf-mutism
C. disturbances in gait
D. patellar hyperreflexia
E. delayed pubertal development

41. All the following conditions are associated with Graves' disease **EXCEPT**

A. type 1 diabetes mellitus
B. vitiligo
C. iron deficiency anemia
D. alopecia areata
E. celiac disease

42. Adverse reaction reported exclusively with propylthiouracil is

A. severe liver disease
B. agranulocytosis
C. lupus-like polyarthritis syndrome
D. glomerulonephritis
E. antineutrophilic cytoplasmic antibody–positive vasculitis

43. A painless nodule in the thyroid or in the neck is the usual presentation of childhood thyroid cancer.
Of the following, the **MOST** common site of distant metastasis is

A. mediastinum
B. long bones
C. skull
D. brain
E. lungs

44. Hypocalcemia is common in neonates between 12 and 72 hr of life, especially in infants with
A. birth asphyxia
B. sepsis
C. exchange transfusion
D. hypomagnesemia
E. hyperbilirubinemia

45. Aplasia or hypoplasia of the parathyroid glands is often associated with the DiGeorge/velocardiofacial syndrome. All the following regarding this syndrome are true **EXCEPT**
A. occurs in 1 in 4,000 newborns
B. caused by a deletion of chromosome 22q11.2
C. hypocalcemia is transitory in the majority
D. hypocalcemia can have its onset later in life
E. conotruncal defects of the heart occur in 75% of cases

46. There is a spectrum of parathyroid deficiencies with clinical manifestations varying from no symptoms to those of complete and longstanding deficiency.
Early manifestations include
A. tingling of the hands and feet
B. muscular pain and cramps
C. carpopedal spasms
D. convulsions
E. delayed irregular teeth eruption

47. Laboratory findings in hypoparathyroidism include the following **EXCEPT**
A. low serum calcium (5-7 mg/dL)
B. elevated serum phosphorus (7-12 mg/dL)
C. elevated serum alkaline phosphatase
D. level of 1,25(OH)2D3 is usually low
E. normal serum magnesium

48. Episodic symptomatic hypocalcemia occurs in the Kenny-Caffey syndrome, the latter is characterized by the following **EXCEPT**
A. medullary stenosis of the long bones

B. short stature
C. delayed closure of the fontanel
D. advanced bone age
E. eye abnormalities

49. Type Ia accounts for the majority of patients with pseudohypoparathyroidism (PHP), they have the following skeletal abnormalities **EXCEPT**
A. brachydactyly
B. dimpling of the dorsum of the hand
C. the three middle fingers are the same length
D. bowing
E. thickening of the calvaria

50. Parathyroid crisis is manifested by the following **EXCEPT**
A. serum calcium levels >15 mg/dL
B. progressive polyuria
C. azotemia
D. stupor
E. coma

51. The **MOST** consistent and characteristic radiographic finding of primary hyperparathyroidism is
A. gross trabeculation of the skull
B. generalized rarefaction
C. cysts
D. resorption of subperiosteal bone
E. radiographic signs of rickets

52. All the following are causes of infantile hypercalcemia **EXCEPT**
A. subcutaneous fat necrosis
B. metaphyseal chondrodysplasia
C. lactase/disaccharidase deficiency
D. blue diaper syndrome
E. proximal renal tubular acidosis

53. Glucocorticoids play a major role in immune regulation. They increase circulating
A. polymorphonuclear cell
B. monocytes
C. eosinophils
D. lymphocytes
E. basophils

54. Glucocorticoids readily penetrate the blood–brain barrier and have direct effects on brain metabolism. They have the following actions **EXCEPT**

A. decrease brain edema
B. insomnia
C. increased irritability
D. impaired memory
E. improved ability to concentrate

55. Patients with mineralocorticoid deficiency can develop the following **EXCEPT**

A. weight loss
B. hypotension
C. metabolic alkalosis
D. hyponatremia
E. hyperkalemia

56. The **MOST** common cause of primary adrenal insufficiency in infancy is

A. congenital adrenal hyperplasia
B. isolated glucocorticoid deficiency
C. X-linked adrenal hypoplasia congenita
D. Lemli-opitz syndrome
E. adrenoleukodystrophy

57. The clinical presentation of adrenal insufficiency depends on the age of the patient, the usual presentation of adrenal insufficiency in infancy is

A. ketosis
B. hyperpigmentation
C. orthostatic hypotension
D. hypoglycemia
E. hypernatremia

58. The **MOST** definitive test for adrenal insufficiency is measurement of

A. blood sugar
B. cortisol before and after administration of ACTH
C. serum sodium
D. arterial blood gases
E. urinary excretion of sodium and chloride

59. Patients with autoimmune Addison disease must be closely observed for the development of other autoimmune disorders.
Of the following, the **MOST** commonly associated disorder in children is

A. alopecia
B. vitiligo

C. chronic active hepatitis
D. type 1 diabetes mellitus
E. hypoparathyroidism

60. High-dose glucocorticoids (the equivalent of >10 times physiologic cortisol secretion) can be administered without requiring a subsequent tapering for
A. 5 day
B. 7 day
C. 10 day
D. 14 day
E. 21 day

61. Classic 21-hydroxylase deficiency occurs in approximately 1 in 15,000- 20,000 births in most populations. Of affected infants, the salt-losing form constitute about
A. 40%
B. 50%
C. 60%
D. 70%
E. 80%

62. The signs and symptoms of congenital adrenal hyperplasia (salt wasting form of the disease) caused by 21-hydroxylase deficiency typically first develop in affected infants at approximately
A. 1 week of age
B. 2 weeks of age
C. 3 weeks of age
D. 4 weeks of age
E. 6 weeks of age

63. Significantly virilized females with congenital adrenal hyperplasia (CAH) usually undergo surgery between
A. 2- 6 mo of age
B. 7-12 mo of age
C. 13-24 mo of age
D. 5-9 year of age
E. 10-14 year of age

64. Cortisol deficiency in congenital adrenal hyperplasia (salt wasting form of the disease) is treated with hydrocortisone
A. 5-10 mg/m2/24 hr
B. 10-15 mg/m2/24 hr
C. 15-20 mg/m2/24 hr

D. 20-25 mg/m2/24 hr
E. 25-30 mg/m2/24 hr

65. In older children with Cushing syndrome, in addition to obesity, a common early manifestation is
A. purplish striae on abdomen
B. short stature
C. hypertension
D. hyperglycemia
E. osteoporosis

66. All the following tests are of diagnostic utility in the diagnosis of Cushing syndrome **EXCEPT**
A. midnight cortisol levels
B. nighttime salivary cortisol levels
C. glucose tolerance test
D. urinary excretion of free cortisol
E. dexamethasone suppression test

67. In primary aldosteronism, one of the following is low
A. serum PH
B. serum carbon dioxide
C. serum sodium
D. serum chloride
E. serum calcium

68. Clinical manifestations of pheochromocytomas include the following **EXCEPT**
A. hypertension
B. convulsions
C. pulmonary edema
D. obesity
E. good appetite

69. Calcification within the adrenal glands may occur in a wide variety of situations, some serious and others of no obvious consequence.
Adrenal calcifications are bilateral in
A. neuroblastomas
B. ganglioneuromas
C. Wolman disease
D. cortical carcinomas
E. pheochromocytomas

70. The major causes of incidentalomas are
A. benign adenomas
B. pheochromocytomas
C. adrenocortical carcinoma
D. hemorrhagic cysts
E. myelolipomas

71. Congenital hypogonadotropic hypogonadism include
A. gonadal dysgenesis
B. Klinefelter syndrome (47,XXY)
C. Noonan syndrome
D. cystic fibrosis
E. Laurence-Moon- Biedl syndrome

72. Acquired hypogonadotropic hypogonadism include
A. chemotherapy
B. malnutrition
C. cystic fibrosis
D. infection (e.g., mumps)
E. infarction (testicular torsion)

73. Klinefelter syndrome is the most common sex chromosomal aneuploidy in males, all the following features are true **EXCEPT**
A. 80% of them have a 47,xxy chromosome complement
B. diagnosis is rarely made before puberty
C. psychiatric disorders may be apparent long before defects in sexual development
D. few complete high school
E. approximately 80% of adults have gynecomastia

74. True gynecomastia is characterized by the presence of a palpable fibroglandular mass (located concentrically beneath the nipple and areolar region) at least
A. 0.5 cm in diameter
B. 1 cm in diameter
C. 1.5 cm in diameter
D. 2 cm in diameter
E. 2.5 cm in diameter

75. Weaker associations with gynecomastia are seen in
A. spironolactone
B. alkylating agents
C. ketoconazole

D. cimetidine
E. opiates

76. Significant gynecomastia is seen in
A. hyperthyroidism
B. Klinefelter syndrome
C. 17-ketosteroid reductase deficiency
D. 11β-hydroxylase deficiency
E. Peutz-Jeghers syndrome

77. All prepubertal gynecomastia cases, as well as pubertal cases with suspicious features, should be investigated.
Of the following, the initial laboratory evaluation should include
A. karyotype
B. dehydroepiandrosterone sulfate
C. liver function tests
D. thyroid function tests
E. renal function tests

78. Many patients with Turner syndrome are recognizable at birth because of
A. loose skin folds at the nape of the neck
B. cubitus valgus
C. webbing of the neck
D. widely spaced nipples
E. hyperconvex fingernails

79. The **MOST** common cardiac defect in Turner syndrome is
A. aortic coarctation
B. isolated nonstenotic bicuspid aortic valves
C. aortic stenosis
D. mitral valve prolapse
E. anomalous pulmonary venous drainage

80. The 47,XXX (trisomy) chromosomal constitution is the most frequent extra-X chromosome abnormality in females, occurring in almost 1 in 1,000 liveborn females.
It is characterized by the following **EXCEPT**
A. maternal meiotic nondisjunction is the most common cause
B. normal female phenotype
C. sexual development and menarche are delayed
D. tall and gangly
E. behavior disorders

81. The **MOST** common cardiac defect in Noonan syndrome is
   A. aortic coarctation
   B. pulmonary valvular stenosis
   C. aortic stenosis
   D. mitral valve prolapse
   E. anomalous pulmonary venous drainage

82. Familial isolated gonadotropin deficiency associated with anosmia is
   A. Noonan syndrome
   B. Bloom syndrome
   C. Werner syndrome
   D. Kallmann syndrome
   E. Prader-Willi

83. Children with type 2 DM often seek medical care because of
   A. weight loss
   B. excessive weight gain
   C. polyuria
   D. polydipsia
   E. anorexia

84. The current criteria for the diagnosis of type 1 and type 2 DM is a fasting blood glucose that exceeds
   A. 110 mg/dL
   B. 115 mg/dL
   C. 120 mg/dL
   D. 125 mg/dL
   E. 130 mg/dL

85. The clearest evidence of a role for viral infection in human type 1 DM is seen in
   A. acquired rubella infection
   B. live-virus rubella immunization
   C. enteroviral infection
   D. congenital rubella syndrome
   E. mumps infection

86. The following factors are implicated in the pathogenesis of type 1 DM **EXCEPT**
   A. prenatal influences
   B. diet in infancy
   C. viral infections
   D. excessive exposure to certain infections
   E. psychologic stress

87. Falsely low HbA_1c levels are noted in the following conditions **EXCEPT**

A. aplastic anemia
B. pure red cell aplasia
C. blood transfusions
D. cirrhosis
E. renal disease treated with erythropoietin

88. Subjects with type 1 DM have evidence for celiac disease in

A. 5-10%
B. 11-15%
C. 16-20%
D. 21-25%
E. 26-30%

89. Diabetic ketoacidosis (DKA) is the end result of the metabolic abnormalities resulting from a severe deficiency of insulin or insulin effectiveness.
DKA is characterized by the following **EXCEPT**

A. ketonuria
B. normal ion gap
C. decreased serum bicarbonate
D. decreased pH
E. elevated effective serum osmolality

90. To continue the insulin infusion in treaterment of DKA without causing hypoglycemia, glucose must be added to the infusion. We typically recommend that glucose be added as a 10% solution when the serum glucose has decreased

A. <100 mg/dL
B. <150 mg/dL
C. <200 mg/dL
D. <250 mg/dL
E. <300 mg/dL

91. Diabetic ketoacidosis (DKA) may be arbitrarily classified as mild, moderate, or severe. Venous pH in severe DKA is

A. <7.05
B. <7.10
C. <7.15
D. <7.20
E. <7.25

92. Any child with diabetic ketoacidosis (DKA) can be easily transitioned to oral intake and subcutaneous insulin when the following criteria are found **EXCEPT**

A. total CO_2 >15 mEq/L
B. pH >7.25
C. sodium stable between 135 and 145 mEq/L
D. no emesis
E. no dehydration

93. Cerebral edema complicating DKA remains the major cause of morbidity and mortality in children and adolescents with T1 DM.
All the following are true **EXCEPT**
A. early bolus administration of insulin is a risk factor
B. high volumes of fluid is a risk factor
C. the incidence of cerebral edema in children with DKA has not changed over the past 15-20 yr
D. radiographic imaging is frequently helpful in making the diagnosis of cerebral edema
E. its etiology remains unknown

94. Nonketotic hyperosmolar coma is characterized by the following **EXCEPT**
A. severe hyperglycemia
B. nonketotic acidosis
C. severe dehydration
D. hypothermia
E. positive babinski signs

95. HbA_1c values may be spuriously elevated in
A. thalassemia
B. sickle cell disease
C. iron deficiency anemia
D. leukemia
E. aplastic anemia

96. Microvascular complications of DM include
A. nephropathy
B. accelerated coronary artery disease
C. cerebrovascular disease
D. peripheral vascular disease
E. peripheral neuropathies

97. Guidelines suggest that prepubertal T1DM patients should commence screening for retinopathy
A. at diagnosis
B. 1 yr after diagnosis

C. 3yr after diagnosis
D. 5 yr after diagnosis
E. 10yr after diagnosis

98. Guidelines suggest that diabetic patients should commence screening for celiac disease
A. at diagnosis
B. 1 yr after diagnosis
C. 3 yr after diagnosis
D. 5 yr after diagnosis
E. 10 yr after diagnosis

99. Transient elevation of urinary albumin can occur in the following conditions **EXCEPT**
A. hypoglycemia
B. strenuous exercise
C. urinary tract infections
D. heart failure
E. acute febrile illness

100. Signs of insulin resistance or conditions associated with insulin resistance in T2 DM in children include the following **EXCEPT**
A. acanthosis nigricans
B. hypertension
C. dyslipidemia
D. increased waist : hip ratio
E. polycystic ovary syndrome

101. The **MOST** commonly used and the only FDA-approved oral agent for the treatment of T2 DM in children and adolescents is
A. thiazolidinediones
B. pramlintide
C. metformin
D. sulfonylureas
E. acarbose

102. Donohue syndrome is characterized by the following **EXCEPT**
A. intrauterine growth restriction
B. fasting hypoglycemia
C. acanthosis nigricans
D. postprandial hyperglycemia
E. profound resistance to insulin

103. One of the following do not appear to be of concern in cystic fibrosis–related diabetes (CFRD)

A. ketoacidosis
B. macrovascular complications
C. microvascular complications
D. frequent infections
E. energy needs

104. When diabetes and thyroid disease coexist, the possibility of autoimmune adrenal insufficiency should be considered. It may be heralded by the followings **EXCEPT**

A. increasing insulin requirements
B. increasing pigmentation of the skin
C. salt craving
D. asthenia
E. postural hypotension

105. Therapy with one of the following drugs usually results in significant insulin resistance leading to glucose intolerance and overt diabetes

A. streptozotocin
B. rodenticide Vacor
C. high-dose oral steroid
D. cyclosporin
E. tacrolimus

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PART XXVI The Endocrine System ANSWERS

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1.**(A)**. Decreased activity of renal α-hydroxylase (which is involved in converting the less-active 25-hydroxyvitamin D into the more active 1,25-$(OH)_2$ D) is a cause in renal insufficiency.
2.**(B)**. Hyponatremia with reduced serum osmolality.
3.**(D)**. Venous pH <7.30.
4.**(C)**. 30%. The percentage is higher in younger children (<5 years old).
5.**(B)**. Prepubertal children generally require about 0.5 U/kg/day. During the middle of puberty, dosages often exceed 1 U/kg/day, whereas postpubertal individuals require 0.75-1.0 U/kg/day. Athletes or those with a low caloric intake may require less insulin. For most children with type 1 diabetes, there are a variety of strategies and combinations of insulin that are currently used. There is a trend toward three or more doses of insulin each day. In addition, more children are on insulin pump therapy.
6.**(B)**. Bone age delayed up to 2-4 years but consistent with height age
7.**(B).** Congenital goiter is seen in only 20% of newborns with congenital hypothyroidism.
8.**(A)**. In children with a solitary nodule, about 30-40% has a carcinoma, 20-30% has an adenoma, and the remainder will have thyroid abscess, thyroid cyst, multinodular goiter, Hashimoto thyroiditis, subacute thyroiditis, or nonthyroidal neck mass.
9.**(C).** The pituitary gland is located at the base of the skull in a saddle-shaped cavity of the sphenoid bone called the sella turcica.
10.**(C).** Sleep, exercise, physical stress, trauma, acute illness, puberty, fasting, and hypoglycemia stimulate the release of GHwhereas hyperglycemia, hypothyroidism, and glucocorticoids inhibit GH release.
11.**(D)**. Arginine vasopressin, oxytocin, angiotensin II, and cholecystokinin stimulate release of CRH and ACTH, whereas atrial natriuretic peptide and opioids inhibit release of CRH and ACTH.
12.**(A)**. Any lesion that damages the hypothalamus, pituitary stalk, or anterior pituitary can cause pituitary hormone deficiency. Because such lesions are not selective, multiple hormonal deficiencies are usually observed. Diabetes insipidus is more frequent in acquired than in congenital hypopituitarism. The most common lesion is the craniopharyngioma.
13.**(C)**. The FDA has approved 8 indications for GH treatment to promote growth. They are GH deficiency, Turner syndrome, chronic renal failure before transplantation, idiopathic short stature, small-for-gestational age short stature, Prader-Willi syndrome, *SHOX* gene abnormality, and Noonan syndrome.

14.**(E)**. Recent studies indicate that GH treatment is associated with a 6-fold increase in the risk for type 2 diabetes and no significant increase in the risk for type 1 diabetes.
15.**(D)**. Regulation of vasopressin (VP) secretion and serum osmolality. Hyperosmolality, hypovolemia, and hypotension are sensed by osmosensors, volume sensors, and barosensors, respectively. These stimulate both VP secretion and thirst. VP, acting on the kidney, causes increased reabsorption of water (antidiuresis). Thirst causes increased water ingestion. The results of these dual negative feedback loops cause a reduction in hyperosmolality or in hypotension or hypovolemia. Additional stimuli for VP secretion include nausea, hypoglycemia, and pain.
16.**(B)**. Acquired NDI can result from hypercalcemia or hypokalemia and is associated with lithium, demeclocycline, foscarnet, clozapine, amphotericin, methicillin, and rifampin.
17.**(A)**. Neonates and young infants are often best treated solely with fluid therapy, given their requirement for large volumes (3 L/m2/24 hr) of nutritive fluid.
18.**(B)**. Cerebral salt wasting appears to be the result of hypersecretion of atrial natriuretic peptide and is seen primarily with central nervous system disorders including brain tumors, head trauma, hydrocephalus, neurosurgery, cerebrovascular accidents, and brain death. Hyponatremia is accompanied by elevated urinary sodium excretion (often >150 mEq/L), excessive urine output, hypovolemia, normal or high uric acid, suppressed vasopressin, and elevated atrial natriuretic peptide concentrations (>20 pmol/L). Thus, it is distinguished from SIADH, in which normal or decreased urine output, euvolemia, only modestly elevated urine sodium concentration, and an elevated vasopressin level occur.
19.**(C).**
20.**(B)**. Precocious puberty is defined by the onset of secondary sexual characteristics before the age of 8 yr in girls and 9 yr in boys.
21.**(C).** Hypothalamic hamartomas are the most common brain lesion causing central precocious puberty.
22.**(E)**. Breast development may regress after 2 yr, often persists for 3-5 yr, and is rarely progressive.
23.**(C)**. Hair appears on the mons and labia majora in girls and perineal and scrotal area in boys; axillary hair generally appears later.
24.**(E)**. Serum TSH levels are the most accurate test of thyroid function. Serum TSH levels are elevated in primary hypothyroidism and suppressed in hyperthyroidism.
25.**(C).** At birth, there is an acute release of TSH; peak serum concentrations reach 60 mU/L 30 min following delivery in full-term infants. A rapid decline occurs in the ensuing 24 hr and a more gradual decline over the next 5 days to <10 mIU/L. The acute increase in TSH produces a dramatic increase in levels of T4 to approximately 16 µg/dL and of T3 to approximately 300 ng/dL in about 4 hr. This T3 seems largely derived from increased peripheral conversion of T4 to T3. T4 levels gradually decrease during the 1st 2 wk of life to 12 µg/dL.

26.**(A)**. TBG levels increase in pregnancy, in the newborn period, with hepatitis, and with administration of estrogens (oral contraceptives), selective estrogen receptor modulators, heroin or methadone, mitotane, 5-fluorouracil, and perphenazine, and they decrease with androgens, anabolic steroids, glucocorticoids, nicotinic acid, and l-asparaginase.
27.**(B)**. Some form of thyroid dysgenesis (aplasia, hypoplasia, or an ectopic gland) is the most common cause of permanent congenital hypothyroidism, accounting for 80-85% of cases.
28.**(C)**. The cord serum T4 is decreased in proportion to gestational age and birthweight. The postnatal TSH surge is reduced, and the more premature, verylow-birthweight infants with complications of prematurity, such as respiratory distress syndrome, actually experience a decrease in serum T4 in the 1st wk of life. As these complications resolve, the serum T4 gradually increases so that generally by 6 wk of life it enters the T4 range seen in term infants.
29.**(D)**.
30.**(D)**. Thyroid-stimulating hormone levels: premature Infants (28-36 wk); 1st wk of life 0.7-27.0 mIU/L; term Infants: birth to 4 days 1.0-17.6 mIU/L; 2-20 wk 0.6-5.6 mIU/L;
5 mo-20 yr 0.5-5.5 mIU/L.
31.**(C)**. Retardation of osseous development can be shown radiographically at birth in approximately 60% of congenitally hypothyroid infants and indicates some deprivation of thyroid hormone during intrauterine life. The distal femoral and proximal tibial epiphyses, normally present at birth, are often absent.
32.**(E)**. The recommended initial starting dose is 10-15 μg/kg/day (totaling 37.5-50.0 μg/ day for most term infants). The starting dose can be tailored to the severity of hypothyroidism. Rapid normalization of thyroid function has been demonstrated to be important in achieving optimal neurodevelopmental outcome.
33.**(B)**. The most common cause of acquired hypothyroidism is chronic lymphocytic (Hashimoto) thyroiditis.
34.**(E)**. Williams syndrome is associated with subclinical hypothyroidism; this does not appear to be autoimmune, as antithyroid antibodies are negative.
35.**(B)**. Children with chronic hepatitis C infectionare at risk for subclinical hypothyroidism; this does not appear to be autoimmune, because antithyroid antibodies are negative.
36.**(C)**. Deceleration of growth is usually the first clinical manifestation, but this sign often goes unrecognized.
37.**(A)**. Sonography is not indicated unless there is a suspicion of a thyroid nodule on neck palpation. In such cases, ultrasound examination is the most accurate study to confirm the presence of a nodule and determine if other, smaller nodules are present. In addition, an ultrasound examination can determine the nodule dimensions, texture (solid vs cystic nature), and presence or absence of other features that might influence a decision to undertake fine-needle aspiration, such as microcalcifications, blurred

margins, “taller-than-wide” shape, intranodular vascular flow, and pathologic-appearing adjacent lymph nodes.
38.**(D)**. The clinical course is variable. The goiter might become smaller or might disappear spontaneously, or it might persist unchanged for years while the patient remains euthyroid.
39.**(D)**.
40.**(E)**. Affected persons are goitrous but euthyroid, have normal pubertal development and adult stature, and have little or no impaired thyroid function.
41.**(C)**. Pernicious anemia.
42.**(A)**. Severe liver disease, including liver failure requiring transplantation, has been reported exclusively with propylthiouracil.
43.**(E)**.
44.**(A)**. Hypocalcemia is common in neonates between 12 and 72 hr of life, especially in premature infants, in infants with asphyxia, and in infants of diabetic mothers (early neonatal hypocalcemia).
45.**(E)**. Associated abnormalities of the third and fourth pharyngeal pouches are common; these include conotruncal defects of the heart in 25%, velopharyngeal insufficiency in 32%, cleft palate in 9%, renal anomalies in 35%, and aplasia of the thymus with severe immunodeficiency in 1%.
46.**(B)**. Muscular pain and cramps are early manifestations; they progress to numbness, stiffness, and tingling of the hands and feet.
47.**(C)**. The serum level of alkaline phosphatase is normal or low.
48.**(D)**. Delayed bone age.
49.**(C)**. The 2nd metacarpal is involved least often. As a result, the index finger occasionally is longer than the middle finger. Likewise, the 2nd metatarsal is only rarely affected.
50.**(B)**. Progressive oliguria.
51.**(D)**. The most consistent and characteristic radiographic finding is resorption of subperiosteal bone, best seen along the margins of the phalanges of the hands.
52.**(E)**. Distal renal tubular acidosis.
53.**(A)**. High doses of glucocorticoids deplete monocytes and lymphocytes, especially T cells. They do so at least in part by inducing cell-cycle arrest in the G1 phase and by activating apoptosis through glucocorticoid receptor–mediated effects. The effects on lymphocytes are primarily exerted on T-helper 1 cells and hence on cellular immunity, whereas the T-helper 2 cells are spared, leading to a predominantly humoral immune response.
54.**(E)**. There is an increase in irritability and emotional lability, with an impairment of memory and ability to concentrate.
55.**(C)**. Patients with mineralocorticoid excess can develop hypertension, hypokalemia, and metabolic alkalosis.
56.**(A)**. The most common causes of adrenocortical insufficiency in infancy are the salt-losing forms of congenital adrenal hyperplasia. Approximately 75% of infants with 21-

hydroxylase deficiency, almost all infants with lipoid adrenal hyperplasia, and most infants with a deficiency of 3β-hydroxysteroid dehydrogenase manifest saltlosing symptoms in the newborn period because they are unable to synthesize either cortisol or aldosterone.

57.**(D)**. Hyperkalemia, hyponatremia, and hypoglycemia are prominent presenting signs of adrenal insufficiency in infants.

58.**(B)**. The most definitive test for adrenal insufficiency is measurement of serum levels of cortisol before and after administration of ACTH; resting levels are low and do not increase normally after administration of ACTH.

59.**(E)**. Patients with autoimmune Addison disease must be closely observed for the development of other autoimmune disorders. In children, hypoparathyroidism is the most commonly associated disorder, and it is suspected if hypocalcemia and elevated phosphate levels are present.

60.**(B)**.

61.**(D)**. Approximately 70% of affected infants have the salt-losing form, whereas 30% have the simple virilizing form of the disorder.

62.**(B)**. Progressive weight loss, anorexia, vomiting, dehydration, weakness, hypotension, hypoglycemia, hyponatremia, and hyperkalemia. Typically first develop in affected infants at approximately 10-14 days of age. Without treatment, shock, cardiac arrhythmias, and death may occur within days or weeks.

63.**(A)**. Significantly virilized females usually undergo surgery between 2-6 mo of age. If there is severe clitoromegaly, the clitoris is reduced in size, with partial excision of the corporal bodies and preservation of the neurovascular bundle; however, moderate clitoromegaly may become much less noticeable even without surgery as the patient grows. Vaginoplasty and correction of the urogenital sinus usually are performed at the time of clitoral surgery; revision in adolescence is often necessary.

64.**(C)**. Cortisol deficiency is treated with glucocorticoids. This often requires larger glucocorticoid doses than are needed in other forms of adrenal insufficiency, typically 15-20 mg/m2/24 hr of hydrocortisone daily administered orally in 3 divided doses. Affected infants usually require dosing at the high end of this range. Double or triple doses are indicated during periods of stress, such as infection or surgery.

65.**(B)**. In older children, in addition to obesity, short stature is a common presenting feature. Gradual onset of obesity and deceleration or cessation of growth may be the only early manifestations.

66.**(C).** The glucose tolerance test is often abnormal but is of no diagnostic utility. Levels of serum electrolytes are usually normal, but potassium may be decreased, especially in patients with tumors that secrete ACTH ectopically.

67.**(D)**. Hypokalemia occurs frequently. Serum pH and the carbon dioxide and sodium concentrations may be elevated and the serum chloride and magnesium levels decreased. Serum levels of calcium are normal, even in children who manifest tetany.

68.**(D)**. Patients have a good appetite but because of the hypermetabolic state may not gain weight, and severe cachexia may develop.

69.**(C)**. Infants with Wolman disease, a rare lipid disorder caused by a deficiency of lysosomal acid lipase, have extensive bilateral calcifications of the adrenal glands.
70.**(A)**. Benign, hormonally inactive adrenocortical adenomas make up the majority of incidentalomas.
71.**(E)**.

- Follicle-stimulating hormone (FSH) and luteinizing hormone (LH) resistance
- Mutations in steroid synthetic pathways
- Gonadal dysgenesis
- Klinefelter syndrome (47,XXY)
- Noonan syndrome (PTPN-11 gene mutation in many cases)
- Cystic fibrosis (infertility)

are congenital causes of hypergonadotropic hypogonadism (primary hypogonadism; testes).
72.**(B)**.

- Anorexia nervosa
- Drug use
- Malnutrition
- Chronic illness, especially Crohn disease
- Hyperprolactinemia
- Pituitary tumors
- Pituitary infarction
- Infiltrative disorders (e.g., histiocytosis, sarcoidosis)
- Hemosiderosis and hemochromatosis
- Radiation

are acquired causes of hypogonadotropic hypogonadism (secondary hypogonadism; hypothalamic-pituitary).
73.**(D)**. Verbal cognitive defects and underachievement in reading, spelling, and mathematics are common. By late adolescence, many boys with Klinefelter syndrome have generalized learning disabilities, most of which are language based. Despite these difficulties, most complete high school.
74.**(A)**.
75.**(E)**. Weaker associations are seen with a large number of other medications and drugs of abuse, including opiates, alcohol, and marijuana, although the association with marijuana may not be as strong as previously thought.
76.**(B)**. Significant gynecomastia is seen in 50% of adolescents with Klinefelter syndrome.
77.**(D)**. No laboratory evaluation is indicated in routine cases with no other associated abnormality but all prepubertal cases, as well as pubertal cases with suspicious features, should be investigated; initial laboratory evaluation should include thyroid function tests (to rule out hyperthyroidism), testosterone, estradiol, human chorionic gonadotropin, luteinizing hormone, and prolactin levels.

78.**(A)**. Many patients with Turner syndrome are recognizable at birth because of a characteristic edema of the dorsa of the hands and feet and loose skinfolds at the nape of the neck. Low birthweight and decreased birth length are common.
79.**(B)**. Regardless of the age, all patients with Turner syndrome at the time of diagnosis need comprehensive cardiovascular evaluation by a cardiologist specializing in congenital heart disease. Complete cardiologic evaluation, including echocardiography, reveals isolated nonstenotic bicuspid aortic valves in one third to one half of patients. In later life, bicuspid aortic valve disease can progress to dilation of the aortic root. Less-frequent defects include aortic coarctation (20%), aortic stenosis, mitral valve prolapse, and anomalous pulmonary venous drainage.
80.**(C)**. Sexual development and menarche are normal. Most pregnancies have resulted in normal infants.
81.**(B)**. The phenotype differs from Turner syndrome in several respects. Cognitive impairment is often present, the cardiac defect is most often pulmonary valvular stenosis or an atrial septal defect rather than an aortic defect, normal sexual maturation usually occurs but is delayed by 2 yr on average, and premature ovarian failure has been reported.
82.**(D)**.
83.**(B)**. The presentation of T2DM is typically more insidious than that with T1DM. In contrast to patients with T1DM who are usually ill at the time of diagnosis and whose presentation rarely spans more than a few weeks, children with T2DM often seek medical care because of excessive weight gain and fatigue as a result of insulin resistance and/or the incidental finding of glycosuria during routine physical examination.
84.**(D)**. It should be noted that a fasting blood glucose thatexceeds 125 mg/dL (6.9 mmol/L) is the accepted criterion for the diagnosis of diabetes.
85.**(D)**. The clearest evidence of a role for viral infection in human T1DM is seen in congenital rubella syndrome. Prenatal infection with rubella is associated with β-cell autoimmunity in up to 70%, with development of T1DM in up to 40% of infected children.
86.**(D)**. A number of factors, including prenatal influences, diet in infancy, viral infections, lack of exposure to certain infections even psychologic stress, are implicated in the pathogenesis of T1DM, but their exact role and the mechanism by which they trigger or aggravate autoimmunity remains uncertain.
87.**(A)**. Falsely low HbA1c levels are noted in hemolytic anemias, pure red cell aplasia, blood transfusions, and anemias associated with hemorrhage, cirrhosis, myelodysplasias, or renal disease treated with erythropoietin.
88.**(A)**. Fifteen to 30% of subjects with T1DM have elevated thyroid-stimulating hormone (TSH) and antithyroid antibodies and close to 5-10% have evidence for celiac disease. These diseases share common genes and likely the same interplay between environmental and immunologic factors.
89.**(B)**. An increased ion gap.

90.**(C).** To continue the insulin infusion without causing hypoglycemia, glucose must be added to the infusion. We typically recommend that glucose be added as a 5% solution when the serum glucose has decreased <300 mg/dL and as a 10% solution when the serum glucose has decreased <200 mg/dL. The insulin infusion can also be lowered from the initial maximal rate if, despite the above outlined interventions, the serum glucose falls further.
91.**(C).**
92.**(B)**. Any child can be easily transitioned to oral intake and subcutaneous insulin when DKA has resolved (total CO2 >15 mEq/L; pH >7.30; sodium stable between 135 and 145 mEq/L; no emesis). The first dose of short acting subcutaneous insulin is given with a meal, and the insulin drip is discontinued approximately 30 min later.
93.**(D)**. Radiographic imaging is frequently unhelpful in making the diagnosis of cerebral edema.
94.**(D)**. This syndrome is characterized by severe hyperglycemia (blood glucose >800 mg/dL), absence of or only slight ketosis, nonketotic acidosis, severe dehydration, depressed sensorium or frank coma, and various neurologic signs that may include grand mal seizures, hyperthermia, hemiparesis, and positive Babinski signs.
95.**(A)**. Depending on the method used for determination, HbA1c values may be spuriously elevated in thalassemia (or other conditions with elevated hemoglobin F) and spuriously lower in sickle cell disease (or other conditions with high red blood cell turnover).
96.**(A)**. Complications of DM can be divided into 3 major categories: (1) microvascular complications, specifically, retinopathy and nephropathy; (2) macrovascular complications, particularly accelerated coronary artery disease, cerebrovascular disease, and peripheral vascular disease; and (3) neuropathies, both peripheral and autonomic, affecting a variety of organs and systems. In addition, cataracts may occur more frequently.
97.**(D)**. Screening for retinopathy commenced after 5 yr duration in prepubertal children, after 2 yr in pubertal children.
98.**(A)**. Screening for thyroid disease and celiac disease commenced at diagnosis.
99.**(A)**. Short-term hyperglycemia, strenuous exercise, urinary tract infections, marked hypertension, heart failure, and acute febrile illness can cause transient elevation urinary albumin excretion. There is marked day-to-day variability in albumin excretion, so at least 2 of 3 collections done in a 3-6 mo period should show elevated levels before microalbuminuria is diagnosed and treatment is started.
100.**(D)**.
101.**(C)**. The most commonly used and the only FDA-approved oral agent for the treatment of T2DM in children and adolescents is metformin. Renal function must be assessed before starting metformin as impaired renal function has been associated with potentially fatal lactic acidosis.
Significant hepatic dysfunction is also a contraindication to metformin use, although mild elevations in liver enzymes may not be an absolute contraindication. The usual

starting dose is 500 mg once daily. This may be increased to a maximum dose of 2,000 mg/day. Abdominal symptoms are common early in the course of treatment, but in most cases they will resolve with time.

102.**(C).** Extreme insulin resistance, acanthosis nigricans, abnormalities of the teeth and nails, and pineal hyperplasia are features of Rabson-Mendenhall syndrome.

103.**(B)**. Macrovascular complications do not appear to be of concern in CFRD, perhaps because of the shortened life span of these patients.

104.**(A)**. Decreasing insulin requirements.

105.**(C).** High-dose oral or parenteral steroid therapy usually results in significant insulin resistance leading to glucose intolerance and overt diabetes. The immunosuppressive agents cyclosporin and tacrolimus are toxic to β cells, causing IDDM in a significant proportion of patients treated with these agents. Their toxicity to pancreatic β cells was 1 of the factors that limited their usefulness in arresting ongoing autoimmune destruction of β cells. Streptozotocin and the rodenticide Vacor are also toxic to β cells, causing diabetes.